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# Centro-temporal benign epilepsy in Saudi children

MOH'D M.U. KABIRAJ, SAAD AL RAJEH, ADNAN AWADA, MOHAMMAD ABDULJABBAR, ABDULKADER DAIF, ABDULRAHMAN AL TAHAN & MUNIRA AL BUNYAN

*Division of Neurology and Clinical Neurophysiology, King Khalid University Hospital—King Saud University, Riyadh, Saudi Arabia*

Correspondence to: Dr. M.M.U. Kabiraj, Assistant Professor, King Khalid University Hospital, P.O. Box 2925, Riyadh 11461 Saudi Arabia

A review of 36 interictal EEGs compatible with the diagnosis of benign childhood epilepsy with centro-temporal spikes (BECTS) was made in this study. These children constituted 31% of the children with focal epileptic EEG activity observed in the same period. The dipolic spikes occurred either as a single focus (78%) or as two independent foci with a greater preponderance to the right (22%). The onset of seizures before the age of 5 years (Group I) is 53% and between 6 and 10 years (Group II) is 47%. Clinical evaluation showed that 60% of the patients in Group I and 75% in Group II had lateralized seizures, although all of them were initially diagnosed as nocturnal generalized tonic–clonic seizures. A follow-up study showed 31% complete recovery, 47% seizure-free under medications, 8% occasional seizures, 8% frequent seizures, and the rest 6% had a single seizure without medication. It is therefore concluded that the syndrome is common in Saudi Arabia and is usually unrecognized by the general physicians. The follow-up of our patients so far confirms the excellent prognosis.

## INTRODUCTION

Benign childhood epilepsy with centro-temporal spikes (BCECTS) is a well-known clinico-electroencephalographic (EEG) syndrome[1–5] and occurs as commonly as childhood absence epilepsy[6]. Interictal electroencephalography shows paroxysmal biphasic spikes or sharp waves, either isolated or followed by slow waves in the centro-temporal regions[7,8]. The syndrome remits before the age of 16 years, most commonly around 9–12 years of age[7–9]. However, this most benign epilepsy remains largely unrecognized or misdiagnosed in Saudi Arabia, probably because most patients, as in our series, initially present as nocturnal generalized tonic–clonic convulsions. Our objective is to raise the level of awareness of BECTS among the primary care physicians in Saudi Arabia by describing the clinical presentations of this common, treatable and reversible syndrome and emphasizing the diagnostic role of interictal EEG.

## MATERIALS AND METHODS

The EEGs of 117 children aged 1–15 years with focal epileptiform activity or focal onset-secondarily generalized discharges were collected from 1985 to 1990 and were reviewed at the Clinical Neurophysiology Laboratory, King Khalid University Hospital (KKUH), Riyadh. These EEGs were recorded according to the International 10-20 System. Recording utilized referential, longitudinal bipolar and transverse bipolar montages. Hyperventilation and photic stimulation were performed on all patients. We considered the records to be compatible with BECTS if they showed the following features: (1) normal background activity, (2) focal biphasic spikes with blunted peak or sharp waves and spike and slow waves which were activated by drowsiness and/or sleep, (3) absence of focal slowing. The foci were identified according to the area of greatest amplitude in the referential montage and where they phase reversed in the bipolar montages.

The medical records of those patients who met these EEG criteria were thoroughly reviewed and the following information was retrieved: age, sex, clinical description and age of onset the seizures, family history, response to treatment and outcome.

## RESULTS

The distribution of the EEG findings of 117 patients selected on an electro-clinical basis,

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showed the following: 36 patients (31%) fulfilled the EEG criteria of BCECTS as defined above. Twenty-two (19%) had complex partial seizures, 26 (22%) had simple partial seizures, 25 (21%) had partial seizures with secondary generalization and 8 (7%) had unclassifiable partial seizures.

Interictal EEGs were done during awake, drowsy and sleep states. The background activity was normal. Topographical distribution of discharges is summarized in Table 1. The biphasic slow spike and sharp waves showed phase reversals independently in the central [Fig. 1(a)], a mid-temporal [Fig. 1(b)], centro-parietal (Fig. 2) and anterior temporal (Fig. 3) regions. Drowsiness and sleep activate these discharges (Fig. 4). Figure 5 shows the bilateral independent foci. A total of 380 randomly occuring epileptiform foci were analysed and a frequency distribution curve was plotted (Fig. 6). Maximum negativity of the foci laid in the centro-temporal regions. (Fig. 6).

Twenty-eight of the 36 patients with EEG suggestive of BCECTS (78%) showed a single EEG focus. Most of these foci were in the central/mid-temporal areas (55%). The rest of the single foci (45%) had a wide range of freedom outside central/centro-temporal areas in both the hemispheres, but most often on the right one (Figs 2 and 3). Table 1 also shows that 45% of the patients had lateralized seizures contra-lateral to the foci, 45% patients could not lateralize and 10% had ipsilateral seizures. Table 2 shows the topographical distribution of eight patients with two independent foci. The EEGs of patients with seizure onset occuring at 6 years of age and above showed foci mainly localized in the central or mid-temporal regions. Table 3 summarizes the clinical characteristics of the seizures of our 36 patients.

The age of onset of seizures ranged from 1 year to 13 years (mean 5.45 years). Those with age of onset of seizures between 1 and 5 years were defined as group I. Those whose seizures began between 6 and 10 years as group II (Table 3). In both the groups, partial seizures constituted the major presentation (67%). The characteristics of the partial seizures were twitching of the angle of mouth, rolling of the eyeball, hemifacial spasm, jerking of the upper extremity and turning of the head to one side. Forty-five per cent of the patients developed speech arrest, i.e. the patients understood spoken words but could not talk (Table 3). One patient, a 9-year-old boy became frightened, grasped people and could not talk during the attack.

Three patients in group I had an early onset of seizures. All three had longer histories of persisting seizures, up to the age of 14 years. Two out of 36 patients had onset of seizures after the age of 10. Both of them had only a single isolated seizure and were not treated with medication. Profuse salivation and abdominal pain were noted in both the groups. A total of 12 patients (33%) (8 from group I and 4 patients from group II) had a nocturnal generalized tonic–clonic seizure (GTCS) (Table 3). Some of them developed post-ictal confusional state, walking aimlessly around the room.

During the follow-up, 11 patients (31%) showed complete recovery. They are now in their adolescence and are seizure-free without medication. Seventeen patients (47%) were seizure-free with medication, although EEG shows epileptiform discharges. Three patients (8%) had infrequent seizures with positive EEG findings. The other three patients had continuing uncontrolled seizures. The last two (6%) had single seizures only and were not treated.

## DISCUSSION

The characteristic EEG patterns of BCECTS syndrome have been widely described[1,4,10]. All our 36 patients were first suspected to have BCECTS on the EEG findings of the characteristics dipolic potentials, their stereotype nature, activated by sleep and their typical topographic

Table 1: Topographical distribution of the EEG discharges with a single focus and lateralized seizures ($n = 28$)

| Topographical areas | Hemisphere | | Lateralized seizures | | |
|---|---|---|---|---|---|
| | Right | Left | Contra | Ipsil | Not descr. |
| Central/Mid-temporal, 15 (54%) | 13 | 2 | 7 | 2 | 6 |
| Parietal, 6 (21%) | 4 | 2 | 3 | 1 | 2 |
| Centro-parietal, 4 (11%) | 1 | 3 | 2 | 0 | 2 |
| Other areas*, 3 (10%) | 1 | 2 | 1 | 0 | 2 |

Contr = contralateral; Ipsil = ipsilateral; Not descr = not described by the patients; *Other areas: tempo-parietal 1; parieto occipital 1; fronto central 1.

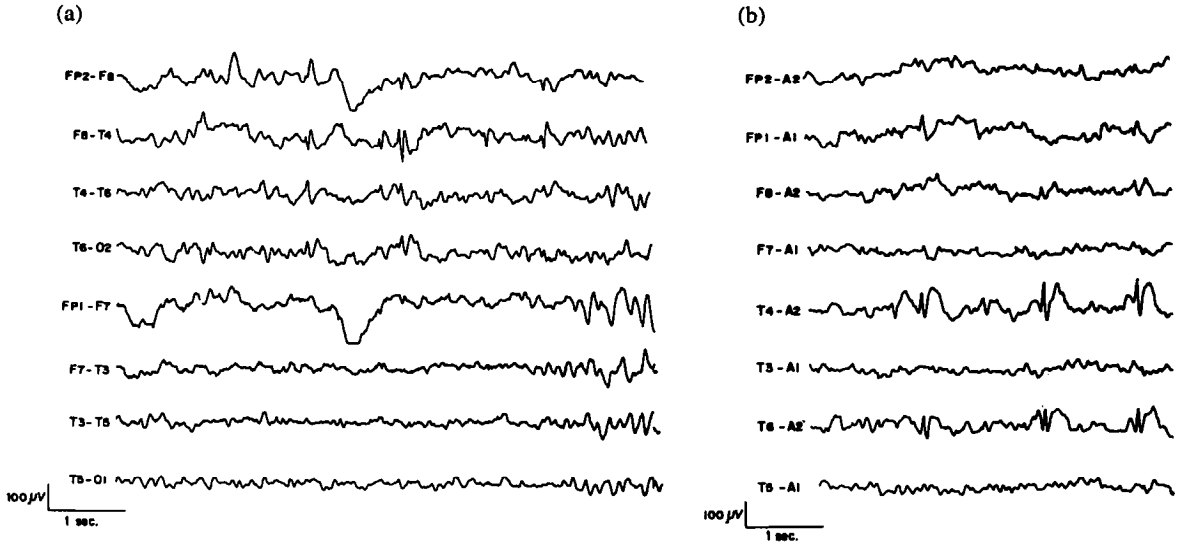


Fig. 1: (a) Electroencephalogram in the state of drowsiness from a 7-year-old boy. Well developed sharp waves are seen in the right, phase reversing in the $T_4$ electrode. (b) Electroencephalogram in the state of drowsiness from a 7-year-old girl. Background is slow. Runs of spikes and slow discharges are seen in the right, mid- and posterior temporal regions.

distributions. Nine of the 20 patients with seizure onset before the age of 6 years had single EEG focal discharges outside the central and mid-temporal areas while 13 of the 16 with a single focus at the central and mid-temporal areas had seizure onset at age of 6 years or after. This suggests, as it was observed by others[2,12] that the epileptogenic foci tend to be scattered around

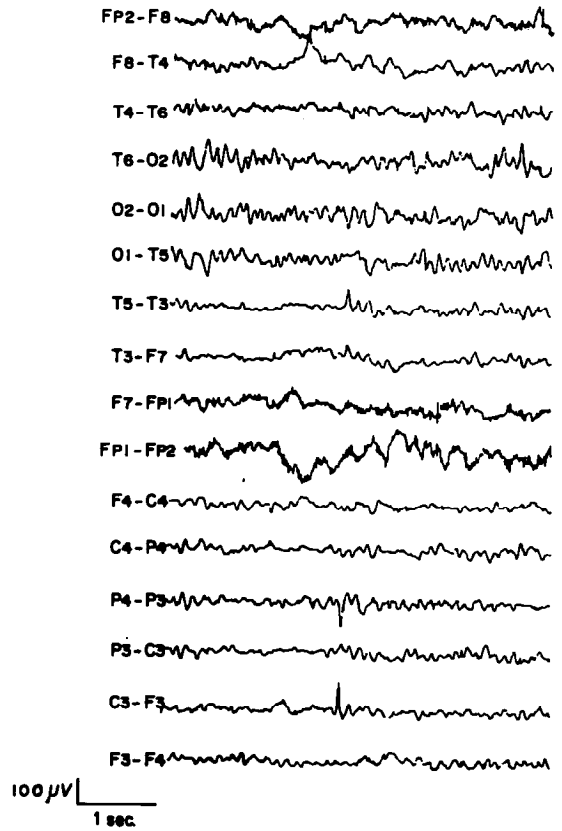


Fig. 2: Electroencephalogram in awakefulness from a 7-year-old boy. Burst of sharp waves seen in the left with an isopotentiality in the left centro-parietal electrodes.

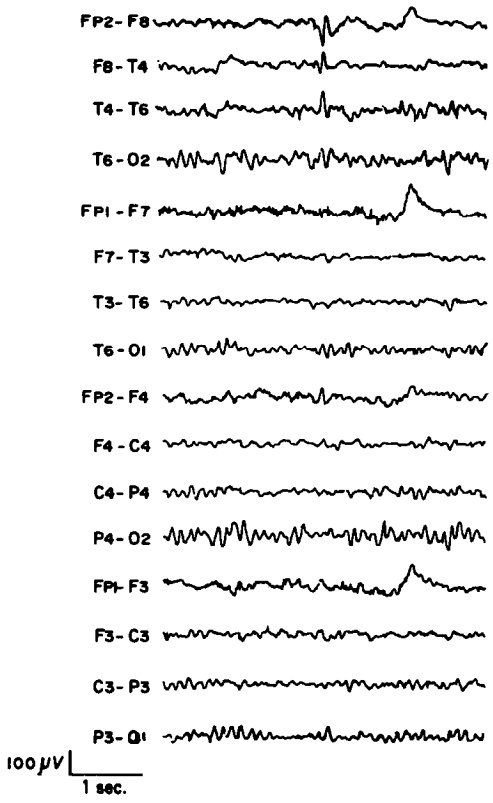


Fig. 3: EEG in wakefulness from a 11-year-old boy. Normal background. A sharp wave focus was seen in the right anterior temporal region.

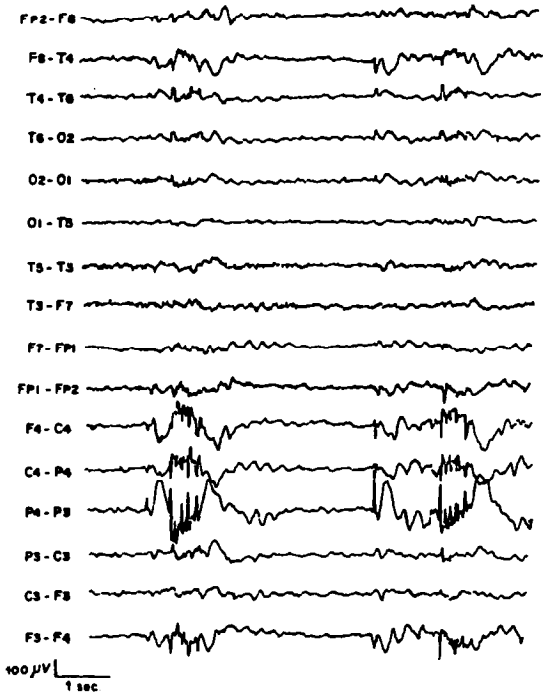


Fig. 4: EEG in sleep state from a 9-year-old boy. The clusters of spikes are seen in the right centro-parietal regions.

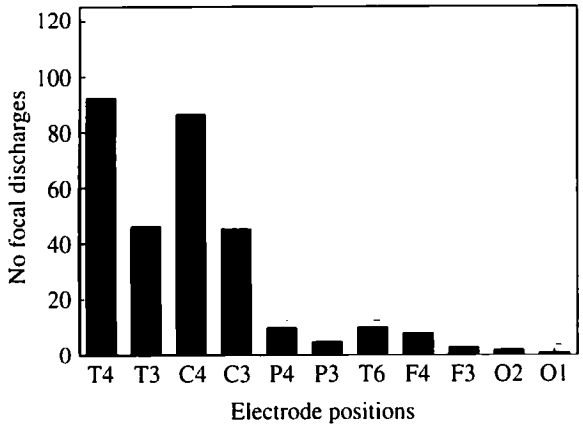


Fig. 6: Frequency-distribution of randomly occuring foci in the different regions of the brain.

the centro-temporal areas before the age of 6, then ultimately migrate to the centro-temporal regions after this age. It is, however, interesting to note that the patients having two independent foci involving both hemispheres did not follow this age-dependent rule of focalization of the syndrome (Table 2).

As noted by Loisequ *et al*[13], the 20 of our 36 patients who had a seizure onset before the age of 6 (Table 3, group I) and had longer seizure duration (mean duration was 4.85 years).

Nocturnal generalized tonic–clonic seizures were not an uncommon initial clinical presentation, occuring in 12 patients (33%) in both groups I and II. EEGs of all of them had single or two independent foci. Many of them were most likely secondarily generalized after focal (partial) onset but the initial partial seizure was probably missed due to the nocturnal occurrence and/or due to inaccurate history[1,5,8].

Although centro-temporal foci usually correlated with contralateral seizures (Table 1), four patients had focal seizure ipsilateral to the EEG discharges. Also seven out of nine patients (78%) with bilateral independent discharges had unilateral partial seizures (Table 2). However, it has

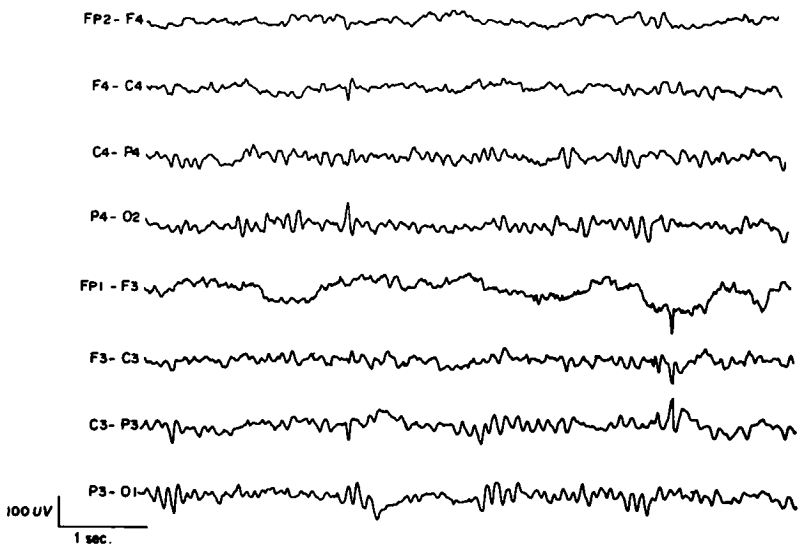


Fig. 5: EEG in drowsiness from a 7-year-old girl, bilateral independent discharges of sharp waves are seen in both the right and left central regions.

Table 2: Topographical distribution of EEG discharges with two individual foci ($n = 18$)

| Age of seizure onset | Sex | Left hemisphere | Right hemisphere |
|---|---|---|---|
| 5 | M | C–MT | C |
| 8 1/2 | M | C–MT | C |
| 6 | M | C | MT |
| 3 | F | C–P | C–P |
| 1 | M | C–MT | C–P |
| 1 1/2 | F | C–MT | C–MT |
| 3 | M | P–O | P–O |
| 2 1/2 | M | P–T | C–M |

C = central; P = parietal; MT = mid-temporal; O = occipital; PT = posterior temporal.

been observed that in BECTS focal epileptiform discharge can be seen in the ipsilateral as well as contralateral hemisphere[14].

Seventeen out of 36 patients (47%) from both the groups developed speech arrest. Patients could understand verbal instructions, but could not talk. Our observations of the preservation of consciousness and speech arrest are consistent with the experience of others[1,3,9]. Nine patients in group I and three patients in group II had autonomic symptoms such as excessive salivation, abdominal pain and appearance of fright as also reported by other investigators[1,4].

The outcome of the syndrome was excellent in our patients. Eleven patients were followed for a mean period of 7.5 years, after the withdrawal of anticonvulsive therapy and found to be seizure-free. Another group of 17 patients was seizure-free with anticonvulsive medications. The serial follow-up EEGs still showed infrequent epileptiform discharges, especially during drowsiness and sleep. They are still attending the epilepsy clinic regularly. When these two groups are combined together, it appears that 28 patients (78%) were seizure-free. The retrospective as well as the prospective studies of others showed complete disappearance of both the clinical manifestations and EEG discharges before reaching adulthood[9,5,15]. As mentioned above, only three patients out of 36 patients (8%) were poorly controlled. They had frequent seizures commencing at 1 to 2.5 years of age. One of them had electroclinical evidence of associated generalized tonic–clonic seizures. Computerized tomography studies were normal. However, it has been reported that in small minority of cases of BCECTS, the seizures were difficult to control[16].

In conclusion, BCECTS appears to be common in Saudi Arabia as in other places but is usually unrecognized by the general physician. Our data support its excellent prognosis and our attitude in managing these patients. We believe that carbamazepine or valproic acid are the drugs of choice in this syndrome and that in the great majority of cases, antiepileptic treatment can be stopped around the age of 16 years or even before this age if the EEG normalizes and the patient becomes seizure free for a few years.

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Table 3: Age of onset of seizures, characters of the seizures and their durations in 36 patients with bects

| Group I ($n = 20$)<br>Age of onset (1–5 years)<br>Mean duration of seizures (4.85) | | Group II ($n = 16$)<br>Age of onset (6–10 years)<br>Mean duration of seizures (1.9 yrs) | |
|---|---|---|---|
| Generalized | 8 (40%) | Generalized | 4 (25%) |
| Partial | 12 (60%) | Partial | 12 (75%) |
| Bucco-lingual/facial | 15 (75%) | Bucco-lingual facial | 13 (81%) |
| Extremities (mainly upper) | 9 (45%) | Extremities (mainly upper) | 8 (50%) |
| Understand, but can't talk | 9 (40%) | Understand/can't talk (frightened) | 8 (50%) |
| Autonomic symptoms | 9 (45%) | Autonomic symptoms | 3 (19%) |

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